Angiographic diagnosis and management of head and neck schwannomas.

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Angiographic Diagnosis and Management of Head and Neck Schwannomas

Schwannomas are tumors derived from nerve sheath cells, which are often located in the head and neck, including the CNS. Although a definitive vascular pattern has been previously characterized for these lesions, preoperative embolization of the more vascular schwannomas has not been described. In a review of eight patients with schwannomas who underwent angiography at our institution since 1987, a characteristic vascular pattern became apparent that helped distinguish these lesions from other lesions of the head and neck. The lesions were moderately vascular with tortuous tumor vessels. Scattered, small puddles of contrast medium seen in the mid-arterial, capillary, and venous phases were believed to be characteristic of these lesions. Multiple feeding vessels were noted in all but one case, but these were only minimally enlarged. No arteriovenous shunting or vascular encasement was identified.

Six of eight lesions were embolized with significant devascularization and no morbidity or mortality.

In patients with head and neck tumors whose angiographic findings include a pattern of moderate hypervascularity, tortuous tumor vessels, and, in particular, scattered contrast puddles without arteriovenous shunting or vascular encasement, schwannoma should be suspected. Embolization is a useful and safe presurgical adjunct in the treatment of vascular schwannomas.


In the era prior to the development and widespread use of cross-sectional imaging techniques, angiography played a major role in the diagnosis and work-up of head and neck tumors [1, 2]. The radiologic and surgical literature contains descriptions of characteristic vascular displacements that helped localize lesions to various compartments (i.e., Meckel cave) [3-6]. Much less is written about characteristic staining patterns of head and neck schwannomas at angiography.

The current radiologic work-up for head and neck schwannomas usually included CT and/or MR imaging. Angiography has been relegated to an ancillary role in those cases in which a highly vascular lesion is suspected (i.e., paraganglioma), critical adjacent arterial structures need to be assessed prior to surgery, or when preoperative embolization is planned. This latter indication is the most common at our institution; six of the eight patients presented here had therapeutic embolization at the time of angiography.

In a review of the eight patients with schwannomas who had angiography at our institution, a particular vascular pattern was noted that helped distinguish schwannomas from other lesions of the head and neck. We will define the vascular pattern seen in our cases and discuss preoperative embolization of these lesions.

Materials and Methods

We retrospectively reviewed the angiograms of eight patients with schwannomas of the head and neck, seven proved histologically and one presumed, performed over a 2-year period. All studies were done on a GE Fluoricon 5000 digital system (General Electric,
Milwaukee, WI). Embolization was performed with a coaxially placed Tracker-18 catheter (Target Therapeutics, San Jose, CA) through a standard 5.5-French diagnostic angiography catheter (Cook, Inc., Bloomington, IN). Polyvinyl alcohol (PVA) particles (Ingenor, Paris, France) were the main embolic material used in all the embolizations; Gelfoam pledgets (Upjohn, Kalamazoo, MI) were occasionally used, either distally for protection of potential anastomoses with internal carotid circulation, proximally to complete the occlusion, or both. In one case of balloon occlusion of the vertebral artery, two #16 Debrun latex Gold Valve balloons (Ingenor, Paris, France) were used, having been detached from a Tracker catheter introduced over a 7-French thin-wall introducer (Balt, Montmorency, France).

The tumor was removed surgically in six cases (1–4, 7, and 8), and the specimens were fixed in 10% buffered formalin and processed routinely with paraffin sections stained with hematoxylin and eosin. The histologic sections from these cases were reviewed to examine microscopic features of the vascular channels. Gross specimens were not available for review.

Results

Angiography was performed in eight cases of head and neck schwannomas at our institution over a 2-year period. Therapeutic embolization was performed in six of the eight cases. In case 1, the schwannoma was located within the left orbital cone and fed by branches of the left ophthalmic artery (Fig. 1). It was thought that this lesion could not be embolized safely.

In seven patients the tumors were surgically removed. In case 6, a 21-year-old woman with a history of right facial numbness and with a dumbbell-shaped lesion in the right Meckel cave, a diagnosis of presumed trigeminal schwannoma was made on the basis of imaging and angiographic findings (Fig. 2). Her 2-year follow-up examination revealed minimal growth of the lesion and no change in neurologic status.
A common angiographic pattern was apparent in these eight cases. Arterial feeders to the lesions were generally of normal caliber or only mildly enlarged. All except the orbital schwannoma had more than one feeding arterial pedicle. The lesions were generally moderately hypervascular with irregular, tortuous tumor vessels. Characteristic small contrast puddles were scattered randomly, appearing in the mid-arterial phase and persisting into the capillary and venous phase (Figs. 3 and 4). There was slight persistence of the capillary stain into the venous phase. No arteriovenous shunting, venous or arterial encasement, or vascular invasion was noted.

In two cases, the lesions appeared smaller at angiography than on cross-sectional imaging studies; presumably, this was due to cystic degeneration (avascular) seen on the imaging studies.

The arterial supply to these lesions was variable, but generally from the external carotid branches as expected by location (Table 1). The orbital schwannoma was supplied exclusively by the ipsilateral ophthalmic artery from the internal carotid artery. In two cases of parapharyngeal space schwannomas (cases 2 and 4), some supply was noted from the ipsilateral vertebral artery. In two cases of Meckel cave seventh nerve schwannomas (cases 5 and 6), some supply from the ipsilateral meningohipophysial trunk was present on internal carotid injections. In case 7, there was sole supply from the left vertebral artery to a C2 neural foramen schwannoma.

Significant devascularization of these lesions was achieved in the six cases in which embolization was performed (Table 1). In all six, the supplying branches of the external carotid artery were superselectively catheterized with Tracker-18 catheters. In four of these cases small PVA particles (150-250 μm) were used. Gelfoam pledgets, usually 2 × 4 mm, were used to protect distal territories, such as the neuromeningeal trunk of the ascending pharyngeal artery, as well as proximally to complete the occlusion of the embolized vessels. In case 3, a schwannoma of low-grade malignancy with extensive supply from branches of the internal maxillary artery, the main internal maxillary trunk was embolized with medium-size PVA particles (250-590 μm) (Fig. 5). Medium-
size PVA particles were also used in case 8, a jugular foramen schwannoma of the vagus nerve.

In case 2, a left parapharyngeal space schwannoma of the vagus nerve, significant vascular supply was noted from the left vertebral artery. Right vertebral angiography revealed a dominant right vertebral system with reflux into the left vertebral artery. Right vertebral angiography revealed a dominant right vertebral system with reflux into the left vertebral artery. A test balloon occlusion of the left vertebral artery at the C1 level, above the level of the tumor feeders, under full systemic heparinization with 5000 units of heparin, was tolerated without ischemic symptoms for 20 min. At this point, the same balloon, filled with iohexol-200 (Winthrop, New York, NY) was detached at C1. The tumor feeders proximal to the balloon were then embolized with small PVA particles and a second, similar balloon was finally detached more proximally at C3–C4 to trap the segment (Fig. 6).

In all cases embolization was tolerated very well without neurologic sequelae. Devascularization of 60–95% of the tumor bed was achieved, in most cases closer to 95%. In five cases in which both embolization and surgery were performed, surgical blood loss was thought to be diminished, presumably due to the preoperative embolization. In case 4, the schwannoma was located at the cervical bifurcation; surgical blood loss was estimated at 25 ml, and the patient was able to be discharged on the second postoperative day. In the case of the low-grade malignant schwannoma with extensive internal maxillary artery supply, there was an estimated 1500 ml blood loss at surgery. Only 60% devascularization was achieved in this case. Additionally, a very extensive surgical procedure was performed in this case, including exploration of the infratemporal fossa and resection of a portion of the mandible that was invaded by tumor. Total operating time was 16 hr, but gross total removal of the lesion was achieved.

Pathology

The six cases in which histologic sections were examined microscopically included four schwannomas (cases 1, 2, 7, and 8), one neurilemmoma, ancient type (case 4), and one malignant schwannoma arising in a neurofibroma (case 3).
TABLE 1: Data and Findings in Eight Patients with Schwannomas of the Head and Neck

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>MR and CT Findings</th>
<th>Angiographic Findings</th>
<th>Estimated Blood Loss During Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>30</td>
<td>M</td>
<td>Left eye proptosis; decreased visual acuity with normal extraocular movement</td>
<td>3 × 2 cm lobulated intracranial left orbital mass that enhanced with contrast; mild bone remodeling</td>
<td>Arterial supply from left ophthalmic artery; no embolization</td>
<td>Minimal</td>
</tr>
<tr>
<td>2</td>
<td>40</td>
<td>F</td>
<td>2-year history of &quot;lump in the throat&quot;</td>
<td>4 × 5 cm low-density left parapharyngeal space mass obliterating left fossa of Rosenmüller, spaying the carotid bifurcation</td>
<td>Arterial supply by left ascending pharyngeal artery, left occipital artery, and muscular branch of left vertebral artery at C2; embolization of ascending pharyngeal and left occipital arteries; left vertebral segment &quot;trapped&quot; with balloons and muscular feeder embolized; 95% devascularized</td>
<td>150 ml</td>
</tr>
<tr>
<td>3</td>
<td>14</td>
<td>M</td>
<td>Several month history of right mandibular pain</td>
<td>5.5 × 6 cm right infratemporal fossa mass of heterogeneous MR signal and CT density distorting posterior maxillary sinus and pterygoid plates, with invasion of right mandible</td>
<td>Multiple arterial feeders including right middle meningeal, right accessory meningeal, right deep temporal, right posterior auricular, and right posterior auricular; right internal maxillary; embolization of right internal maxillary and right posterior auricular; 60% devascularization; right internal carotid test occlusion also performed</td>
<td>1500 ml</td>
</tr>
<tr>
<td>4</td>
<td>24</td>
<td>M</td>
<td>2-year history or left neck swelling</td>
<td>4 × 3 cm heterogeneously dense mass in left posterior styloid, parapharyngeal space, with central low density and thick rim</td>
<td>Arterial supply by left ascending pharyngeal and small muscular branch of left vertebral; embolization of left ascending pharyngeal; 95% devascularization</td>
<td>25 ml</td>
</tr>
<tr>
<td>5</td>
<td>20</td>
<td>F</td>
<td>6-month history of severe headaches, difficulty swallowing, and left facial numbness</td>
<td>4 × 6 cm dumbbell-shaped mass in left Meckel cave with posterior fossa extension compressing the brainstem causing obstructive hydrocephalus</td>
<td>Arterial supply by left accessory meningeal, left middle meningeal, and left meningohypophyseal trunk; embolization of left accessory meningeal and left middle meningeal; 80% devascularization</td>
<td>1500 ml</td>
</tr>
<tr>
<td>6</td>
<td>21</td>
<td>F</td>
<td>1-year history of right facial numbness; history of SLE</td>
<td>2 × 3 cm contrast-enhancing mass involving right Meckel cave with anterior and posterior extension; marked right masticator muscle atrophy</td>
<td>Arterial supply by right middle meningeal, right accessory meningeal, and right meningohypophyseal trunk; embolization of right middle meningeal and right accessory meningeal; 80% devascularization</td>
<td>No surgery or biopsy; 2-year follow-up</td>
</tr>
<tr>
<td>7</td>
<td>10</td>
<td>F</td>
<td>1-month history of right arm and leg weakness; 2-week history of left arm spasm</td>
<td>3 × 3 cm mass in left C2 neural foramen with marked cord compression</td>
<td>Arterial supply from muscular branches of left vertebral; no embolization</td>
<td>250 ml</td>
</tr>
<tr>
<td>8</td>
<td>28</td>
<td>F</td>
<td>High-pitched tinnitus in right ear; difficulty swallowing for 3 months</td>
<td>3 × 3 cm enhancing mass in right jugular foramen</td>
<td>98% of arterial supply from neuromeningeal trunk of right ascending pharyngeal, 1–2% from right AICA; neuromeningeal trunk completely embolized; 98% devascularization</td>
<td>350 ml</td>
</tr>
</tbody>
</table>

Note.—SLE = systemic lupus erythematosus, AICA = anterior inferior cerebellar artery.

The schwannomas exhibited characteristic features of an encapsulated tumor composed of spindle-shaped cells arranged in cellular Antoni A areas, hypocellular myxoid Antoni B areas, and occasional Verocay bodies. The neurilemmoma, ancient type, was similar except for marked hyalinization. The malignant schwannoma arising in a neurofibroma showed areas of marked nuclear pleomorphism, hyperchromaticity, and mitotic figures.

Tumor vessels were usually numerous with expanded sinusoidal-appearing spaces. The majority of these vessels had thin walls. Occasional vessels were present singly or in clusters with hyalinized walls. Most vessels were less than 1 mm in diameter, and only a few were 1–2 mm in diameter. No regions of necrosis or large areas of hemmorhage were present. There were a few small foci of perivascular extravasated erythrocytes and rare foci of hemosiderin deposition. Several vessels of varying sizes showed thrombus formation. Isolated vessels contained PVA or Gelfoam, sometimes with thrombus or inflammatory cells or both. A representative section including some larger vessels is shown in Figure 3.
Variation in the size of vessels among the cases was most apparent with the neurilemmoma, ancient type, which contained numerous larger thin-walled vessels, and with the malignant schwannoma arising in a neurofibroma, which had relatively fewer smaller-size vessels.

Discussion

Schwannomas are most often solitary lesions that arise from the neural sheath of peripheral, spinal, or cranial nerves [7]. They are usually benign but in rare instances can be malignant, the latter occurring de novo or following radiation therapy for benign lesions.

The terminology used to classify nerve sheath tumors has been quite confusing. In the past, schwannomas have been alternatively called neurilemmomas, neuromas, neurinomas, and perineural fibroblastomas [8, 9]. Additionally, some authors [7, 9] have used the term schwannoma to encompass neurilemmoma and neurofibroma, both of which are composed of Schwann cells to some degree. The current literature, however, distinguishes between neurilemmoma (schwannoma) and neurofibroma on a histologic bases [9]. Neither of these lesions should be confused with neuromas. The latter represents an exaggerated repair response to neuronal injury in which a tangle of regenerating axons, fibrous tissue, and Schwann cells form at the site of a severed nerve.

Schwannomas are relatively uncommon lesions; the head and neck, including the CNS, is by far their most common location. In early series, neurofibromas were included with schwannomas, leading to inaccurate estimates of the rate of occurrence of the latter. However, in one large series of 389 schwannomas reported by Gore et al. [10], in which neurofibromas were excluded, 65% of schwannomas were found to be in the CNS with another 13% outside the CNS in the head and neck. Inside the CNS, they most commonly arise from the eighth cranial nerve, followed by spinal nerves and the fifth cranial nerve in descending order of frequency [8]. In the head and neck (outside the CNS) they can occur in any anatomic site, but most frequently in the lateral cervical region.
Schwannomas most often occur in women in their third and fourth decades of life, but may occur at any age. Five of the eight patients reported here are female, ranging in age from 10 to 40 years old. Presentation can be quite variable depending on the size of the lesion, its location, the specific nerve involved, and the presence of anaplasia (malignancy).

Microscopic examination of these cases revealed a vascular pattern dominated by numerous, predominantly thin-walled sinusoidal spaces, in keeping with previously described cases [11–13], and in contrast to angiographic findings in peripheral neurilemmomas.

Angiographic findings revealed that the contrast puddles ranged in size from 1 mm to 4 mm in diameter and were distributed throughout the tumor mass in small clusters. Histologic sections showed only a few larger vessels within this size range. This incongruity between angiographic and histologic findings may be due to the small size and random selection of the tissue specimens from these tumors, which were obtained without angiographic localization. Also, vessel size as seen in histologic sections may appear smaller than angiographic measurements owing to formalin fixation of tissue and lack of vascular pressure.

CT and MR are the prime imaging techniques used in the radiologic work-up of these lesions [14, 15]. Angiography is no longer necessary to diagnose a mass in the head and neck but may be of help in distinguishing between lesions known to occur in certain locations. Additionally, with the ongoing development of interventional techniques in the head and neck, preoperative embolization can be performed on most of these lesions.

A typical angiographic pattern of schwannomas is not clearly established. The radiologic and surgical literature contains few references to the angiography of schwannomas, particularly since the advent of cross-sectional imaging techniques. Several case reports and small series make note of vascular displacements and the presence of tumor "blush" [2–6, 13–18]; in particular, Moscow and Newton [2] as well as Silver et al. [15] described the same characteristic puddles of contrast material at angiography as we have. The presence of prominent capsular veins in eighth nerve tumors has also been described [1]. In one series of 32 patients with "neurinomas" 22 were found to be hypervascular; however, some were considered to be indistinguishable angiographically from meningiomas. These authors described five cases as having irregular tortuous vessels and puddling of contrast material, as we found in our cases. It is unclear from that report whether "neurinoma" refers to schwannoma, neurofibroma, or both. Taversas and Ferrucchi's text [19] claims that schwannomas are hypovascular on angiography. This was also noted in another series of 14 trigeminal neurilemmomas (schwannomas) in which eight were said to be avascular [16]. A pattern of nonshunting hypervascularity with tortuous tumor vessels and, in particular, contrast puddles was noted in each of the eight cases we have presented, and was believed to be characteristic. In our single case of malignant schwannoma, we were unable (even retrospectively) to observe distinguishing features that would have enabled us to predict the histology; this was also the experience of Moscow and Newton [2], who described "no correlation between angiographic appearance of tumor vascularity within intracranial neurinomas and degree of malignancy."

Embolization would have no place in the treatment of avascular tumors; however, the hypervascularity of these schwannomas made them quite suitable for embolization. As with meningiomas, one may argue that these lesions could be operated on without prior embolization. In our experience, this varies from institution to institution. Our surgeons felt that the preoperative embolization had greatly contributed to the safety and efficacy of their procedure. The orbital lesion was the least vascular and also the least amenable to embolization on an anatomic basis (ophthalmic artery supply). In five of the six cases embolized, 80% or greater devascularization was achieved. PVA particles were the chosen embolic agents for obliterating the vascular bed of the tumor while avoiding embolization of potential anastomoses with internal carotid circulation. This resulted in uniformly good outcomes without neurologic sequelae, as well as a reduction in the amount of blood lost during surgery.

In cases of head and neck tumors that are examined angiographically, a pattern of moderate hypervascularity, irregular tumor vessels, and, in particular, scattered contrast puddles and lack of arteriovenous shunting should bring to mind the possibility of schwannoma. These lesions are safely amenable to preoperative embolization in cases in which the surgeon judges that this devascularization will enhance the efficacy and safety of the surgery.

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